

THE EFFECT OF INBREEDING ON EARLY CHILDHOOD MORTALITY: TWELVE GENERATIONS OF AN AMISH SETTLEMENT*

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An unresolved issue in research on child survival is the extent to which familial mortality risk in infancy is due to biological influences net of sociodemographic and economic factors. We examine the effect of consanguinity on early childhood mortality in an Old Order Amish settlement by using the inbreeding coefficient, an explicit measure of the degree of relatedness in one's ancestry. Inbreeding has a net positive effect on neonatal and postneonatal deaths. We find social, demographic, and population-based socio-cultural explanations for this effect among the Amish population, which is known to experience certain genetically transmitted defects associated with mortality.

An unresolved issue in research on child survival is the extent to which familial mortality risk in infancy is due to biological influences net of sociodemographic and economic factors, such as household economic status, household health-related knowledge and attitudes, parental competence in child rearing, and genetic viability (Guo 1993:17; see also Curtis, Diamond, and McDonald 1993; Curtis and Steele 1994; Zenger 1993). One potentially important indicator of genetic viability is degree of consanguinity associated with rare recessive phenotypes (Castilla and Adams 1990; Freire-Maia and Elisbao 1984; Thompson 1990) that affect mortality and morbidity.

We extend prior research on familial mortality risk in the first year of life by using the inbreeding coefficient to predict perinatal, neonatal, and postneonatal mortality. The inbreeding coefficient is an explicit measure of genetic viability unavailable in most data sets. We use data for a population with substantial ancestral inbreeding and include selected sociodemographic and economic measures.

UNOBSERVED MEASURES AND RECENT FAMILIAL MORTALITY RESEARCH

Research on familial mortality risk has reached mixed conclusions when examining whether unmeasured variables produce biased estimates. Curtis et al. (1993) find highly significant variation in their study of familial postneonatal mortality in Brazil, with the effects of birth interval and other

mortality covariates differing little between random-effects and standard logistic models. Guo (1993), studying child mortality in several Latino communities in Guatemala, compares results from hazard models incorporating random family effects with results from a standard hazard model. He finds that genetic factors (as unobserved measures) play a relatively minor role in child survival. Guo concludes that bias from unobserved mortality determinants is likely to be small, although such factors could be of greater importance in more highly consanguineous communities. Zenger (1993) argues that statistical models provide varying conclusions about siblings' mortality risks because the models differ in the way they account for familial association. In her research based on data from Bangladesh, Zenger finds differential results when the preceding child's survival status instead of a random intercept is included in estimation models. Zenger also detects a stronger association for neonatal deaths between immediate pairs of siblings than for those born farther apart. Although models based on random family effects can inform us about the risk of heterogeneity from unobserved variables, explicit measures permit direct examination.

Inbreeding and Familial Mortality Risk

Inbreeding occurs when paired mates share a common ancestor (Crow 1986). The inbreeding coefficient represents the probability that an individual will receive a gene from each parent that is a copy of a single ancestral gene. If parents do not share a common ancestor, the inbreeding coefficient is zero. An inbreeding coefficient of 1.0 indicates a 100% chance that an individual will receive two copies of an ancestral gene. Humans cannot achieve this maximum level of inbreeding, and inbreeding generally is not a major source of mortality in large populations (e.g., the general population of the United States).¹ In certain isolate populations, however, inbreeding levels can be equivalent to mating between close relatives (i.e., brother-sister, uncle-niece).

Most genetic disorders (e.g., inborn errors of metabolism) that are associated with childhood morbidity and mortality are the product of genes that act in an autosomal recessive fashion. Individuals who have only one copy of the defective gene are carriers; that is, they typically do not show any symptoms from the defective gene. For the disorder to manifest itself, there must be two copies (one from the fa-

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1. Alternative explanations have been posited for disorders associated with inbreeding, such as selection and gene pool transitions (e.g., Harper and Roberts 1988; Jakobi and Darlu 1988).

ther and one from the mother) of a defective gene. In the general population, genetic disorders are extremely rare, as both parents typically must be carriers for their child to be born with an autosomal recessive condition.

Using an example outlined by Crow (1986), we can examine the pattern of incidence of a rare recessive disorder that occurs in the general population at an incidence rate of $P_{aa} = 1/8,100$. Using Hardy-Weinberg proportions, we know that $P_{aa} = p_a^2$, where p_a is the gene frequency of the recessive allele. In this example, p_a is $1/90$. Inbreeding increases the incidence of affected individuals in the population. In a mating between first cousins, for example, the inbreeding coefficient is $F = 1/16$. Contrast this with mating between unrelated individuals, which would have an inbreeding coefficient of $F = 0$. The probability of having two copies of the deleterious gene in a mating is given by $P_{aa} = p_a F + p_a^2(1 - F)$. So in an unrelated mating, the equation becomes $P_{aa} = p_a \times 0 + p_a^2(1 - 0) = (1/90)^2 = 1/8,100$, the population frequency. In mating of first cousins, however, the equation becomes $P_{aa} = p_a \times (1/16) + p_a^2(1 - 1/16) = (1/90) \times (1/16) + (1/8,100) \times (15/16) = 6.5625/8,100$.

This example illustrates that in populations such as the Amish, for which inbreeding is relatively high, there may be relatively high instances of disorders that are vanishingly rare in the general population. Because most recessive disorders develop in childhood and typically are life threatening, it is critical to include a measure of inbreeding in an analysis of childhood mortality in consanguineous populations.

The effects of consanguinity have been studied in populations in South Asia (e.g., Bittles 1990; Bittles, Shami, and Rao 1992; Bunday 1992; Reddy and Modell 1995; Shami, Schmitt, and Bittles 1989), Scandinavia (Jorde and Pitkanen 1991; Jorde et al. 1992; Lynch and Greenhouse 1994), Latin America (Castilla and Adams 1990), and certain U.S. subcultures such as the Old Order Amish (Khouri and Cohen 1987; Khouri et al. 1987; Khouri et al. 1987a, 1987b; McKusick et al. 1978). Worldwide reviews of research on the effects of inbreeding in human populations report mixed results (Bittles 1990; Bittles and Makov 1988). Bittles and Makov (1988:164) conclude that the risks to offspring from inbred unions are generally within the limits of acceptability. Some research reports no effects of inbreeding on perinatal and neonatal losses (Al-Awadi et al. 1986) or on infant and child mortality (Reddy and Modell 1995). Other studies find that the effect of inbreeding depends on the period following birth and the community of residence, with consanguinity-related deaths occurring in the neonatal, infant, and childhood periods of more inbred families (Shami et al. 1989).

Most consanguineous populations have high background levels of morbidity and mortality resulting from infectious diseases and poor nutrition, which are associated with low socioeconomic status (Bittles et al. 1992:113; Shami et al. 1989). Yet, not all studies of consanguineous families include socioeconomic and other measures or report tests of statistical significance (see Bittles and Makov 1988) because of data limitations (e.g., Al-Awadi et al. 1986; Reddy and

Modell 1995; Shami et al. 1989). Further, some researchers have observed a positive association between inbreeding and socioeconomic status (Jorde and Pitkanen 1991; Reddy 1987), whereas others have found a negative association (Bittles et al. 1992; Freire-Maia and Elisbao 1984; Tuncbilek and Koc 1994).

There may also be statistical interactions between inbreeding and mortality determinants: Inbreeding might exaggerate the effects of socioeconomic and demographic measures, or socioeconomic and demographic variables might exaggerate the effect of inbreeding. For example, a weak baby might be at increased risk of death in more traditional areas where medical care is less valued or available. The effect of inbreeding may also be mediated by proximate determinants of infant mortality. Highly inbred families might have shorter birth intervals due to the death of a preceding sibling (see Reddy and Modell 1995:244). Consanguineous families might also be larger due to longer reproductive periods (e.g., Reddy 1987; Reddy and Modell 1995; Tuncbilek and Koc 1994), and enhanced fertility may reflect biological or social compatibility (Shami et al. 1989). Estimating the effects of inbreeding within a consanguineous population can help to clarify whether familial mortality risk contributes to the interrelationships between reproductive patterns and child mortality.

Inbreeding and the Old Order Amish

The Old Order Amish is a traditional agricultural religious group that maintains a high degree of isolation from the rest of the community. They are relatively well-off financially and are homogeneous on education, occupation by gender, age at marriage, religion/ethnicity, and, to a lesser extent, health care.² Potentially important social and demographic sources of variability in the Amish community are church district of residence, father's occupation, and family size. In the Lancaster settlement, members of southern church districts are more likely than those in the northern districts to operate farms, have less contact with the surrounding community, live farther from medical facilities, and maintain more traditional values and practices, including interest in larger families (Kraybill 1989).³

2. Amish education ends in the eighth grade and, in the Lancaster settlement, typically is conducted in Amish schools (Ericksen et al. 1979; Kraybill 1989). Few Amish women marry before age 19, and children are rarely born out of marriage. Amish women do not work outside the home after marriage, although they help husbands in the field and some make and sell items to tourists from shops in or adjacent to their homes (Kraybill 1989; D.B. Kraybill, personal communication, February 1992). The Amish do not purchase health insurance, but larger debts incurred for medical care are paid by "Amish church aid" or by the church district (Kraybill 1989:89).

3. In the Amish community, church district of residence has important influences on health behaviors associated with child outcomes. For example, although local nurse-midwives and a physician to the Lancaster, Pennsylvania Amish that we contacted report that most Amish women don't want prenatal care, some are more reluctant than others. The more reluctant women live in the southern church districts of Lancaster County, where farming and traditional lifestyles have been maintained more than in the northern districts (Kraybill 1989; D.B. Kraybill, personal communication, February 1992 and April 1993).

Detailed genealogy records dating to the 1700s have been kept by the Lancaster Amish, providing a well-documented pedigree for up to 12 generations (Beiler 1988). The Amish strongly discourage marriage between close cousins (D.B. Kraybill, personal communication, April 1998). Khoury et al. (1987b) describe the Lancaster Amish genealogy as mutational, with inbreeding from multiple, remote connections rather than from close consanguinity (first-cousin marriages). Certain genetically transmitted defects that are associated with infant mortality have been identified in Amish settlements. An example of a relatively highly recessive genetic disorder in the Lancaster settlement is Ellis-van Creveld syndrome, which is associated with dwarfism of extremities and, in about half the cases, single atrium heart malformation (McKusick et al. 1978:53–54). Perinatal problems and congenital malformations reportedly account for nearly all deaths before one year of age in this settlement (Khoury et al. 1987b:476).

Estimation Model

In the Amish community, education ends at the eighth grade and therefore is controlled by the design of our study. We retain variables for church district of residence and child's date of birth, which represent traditional culture, including interest in and access to health care. We incorporate measures for maternal age, preceding birth interval, birth order, child's gender, and survival status of the immediately preceding sibling.

In preliminary models, we included occupation of the father as a measure of family socioeconomic status. This measure was not statistically significant in any of our models, and we exclude it from the analyses reported in this paper. For perinatal and postneonatal outcomes, we replicated as closely as possible the product terms from the Curtis et al. (1993) study for (1) sibling death, birth interval, and region and (2) maternal age with gender. We also estimated inbreeding interactions with all variables in our study because inbreeding might interact with mortality determinants by exaggerating the effects of socioeconomic and demographic measures, or socioeconomic and demographic variables might exaggerate the effect of inbreeding. We found only one statistically significant interaction (survival of the preceding child by birth interval of 24+ months), and only for perinatal death. Therefore, we report only main effects.

DATA AND METHOD

Our data are derived from the member directory of the Old Order Amish of the Lancaster County, Pennsylvania Settlement (*Amish Directory* 1988; *The Lancaster County Family Register* 1988). The directory represents membership and family records collected and printed by the Lancaster Amish for their internal use. The records include data on births, deaths, marriage dates, residence and church district, and family of origin for nearly every member of the Lancaster Settlement.⁴ Poor recall of marriage patterns can limit avail-

able data in some studies to as few as two generations (Reddy and Modell 1995). Limited genealogical information may be especially problematic in research on populations with comparatively complex pedigrees (see Khoury et al. 1987).

We include all reported deaths in the first year of life and use a simple random sample of about 7% of all surviving singletons (total $n = 2,013$). After we eliminate multiple births and cases with missing values on any variable, the sample size is 1,777.⁵

We analyze the data using logistic regression, which provides coefficients of the log of the expected odds of death (Aldrich and Nelson 1984; Hosmer and Lemeshow 1989). The dependent variables for the three logistic regression models are the three dummy variables representing mortality components: (1) perinatal deaths, infants who were live born but died in the first seven days of life; (2) neonatal deaths, infants who survived the first seven days but died in next three weeks; and (3) postneonatal deaths, infants who survived one month but died before their first birthday.

Coefficient of Inbreeding

A key independent variable in the present study is the inbreeding coefficient, which was calculated using the program KINSHIP (Boyce 1988). If we consider a pedigree with a common ancestor, we can calculate the joint probability that the individual has two copies of the same allele from the common ancestor. The formula that is used for this calculation can be generalized for each common ancestor to

$$F_i = \Sigma (1/2)^n \times (1 + F_a),$$

where n is the number of individuals in a given path between the ancestor and the offspring, F_a is the inbreeding coefficient of the ancestor, if applicable, and these components are summed over all possible paths between the individual and common ancestor (Falconer 1989:87).

We produced the inbreeding coefficients using information from the *Fisher Family History* (Beiler 1988), which documents genealogy relationships for descendants of Christian Fisher, one of the founders of the Old Order Amish community in the United States. We used all members of the Fisher genealogy born between 1757 and 1988 ($N = 52,450$) to calculate the inbreeding coefficient. Although early coefficients (before 1850) are based on single family genealogy, more than 90% of the Amish population is included after 1900. Because inbreeding was not calculated for other founders of this population, the inbreeding coefficients may be underestimated. An inbreeding coefficient could not be calculated for 3.8% of the sample.⁶

5. Because all infant deaths are included in the sample, the sampling weight for infant deaths is 1.0. To simplify the sampling procedure for the large number of live births, we chose pages in the Amish directory as the sampling frames. We calculated the sampling weight for live births as the ratio of the count of selected live births on each page to the total births on that page (using nontwin counts).

6. Each inbreeding coefficient calculated from the Fisher data was matched to its respective case in the *Amish Directory* by date of birth

4. Although a few families or individuals move outside the settlement, which could lead to reporting errors, Kraybill (1989:17) notes that Lancaster Amish migration has never been en masse and in recent years has ended.

We use a continuous measure of inbreeding, which includes inbreeding levels of ancestors (F_a). Other researchers have categorized inbreeding coefficients into "relationship levels" (e.g., McDonald and Diamond 1983). For example, an inbreeding coefficient of 0.25 is equivalent to a brother-sister mating. We base categories of inbreeding on an exponential change with the number of links in the path relating the two individuals, derived from the basic formula of $(1/2)^n$, where n is the number of paths of relationship connecting the parents to a common ancestor. In small pedigrees (with only one path of inbreeding), this level can be readily categorized into relationship levels. Because the inbreeding levels in the Amish data include substantial ancestral inbreeding, discrimination between inbreeding levels is not as clear as in less complex pedigrees. Consequently, categorizing the data into relationship levels in this genealogy would result in inaccurate representation and loss of information (see also Khoury et al. 1987b).

Other Independent Variables

Other independent measures of the present study include church district, which is operationalized as northern (less traditional), southern (more traditional), and other (outside Lancaster County) districts. Maternal age is calculated as the difference between the index child's date of birth and the mother's date of birth. Birth order is defined as the number of prior live-born siblings of the index child, plus one. We include a dummy variable for first births: Deleterious outcomes may affect reproductive strategies, and first births are more prone to deleterious outcomes from nongenetic factors.⁷ Birth interval is operationalized as the time between the preceding sibling's date of birth and the index child's date of birth and is collapsed into three categories: (1) less than 18 months, (2) 18–23 months and (3) 24 months or longer. A dummy variable for neonatal sibling death represents survival status of the immediately preceding sibling. We also include gender of the index child and birth year, which represents variation over time in norms associated with health and reproductive care. Because of the homogenous character of the Amish population, we control for cultural and normative variables that are unmeasured (and even unknown) in most research, including those associated with socioeconomic and marital status, religion/ethnicity, and, to a lesser extent, health care.

Some studies report that birth weight operates as an intervening variable on infant survival, as do intrauterine growth rate, birth complications, and prenatal care. No information is available in the Amish records for these variables. None of these variables, however, are antecedent to any of the independent variables we report. Hence, absence of these proximate causes from our models does not influ-

(mm,dd,yy) and death (if applicable) of the index child, gender of the index child, and both parents' dates of birth and death. When only one digit in one of the five or six dates did not match, we assumed it was a transcription error and accepted the match.

7. Because a first-born child is a special case, we checked for possible interactions between inbreeding and first-born status. We found none.

TABLE 1. WEIGHTED DESCRIPTIVE STATISTICS FOR VARIABLES USED IN THE ANALYSES*

Variable	Mean	Standard Deviation	Minimum	Maximum
Inbreeding Coefficient	0.0166	0.0097	0.0000	0.0602
Birth Year	1968.66	15.64	1917	1988
Female (1 = yes)	0.46	0.50	0.00	1.00
Church District of Residence				
Northern	0.51	0.50	0.00	1.00
Southern	0.30	0.46	0.00	1.00
Other	0.18	0.38	0.00	1.00
Maternal Age				
< 21 years	0.10	0.30	0.00	1.00
21–34 years	0.75	0.44	0.00	1.00
35+ years	0.15	0.36	0.00	1.00
First-Born Child	0.20	0.40	0.00	1.00
Birth Order	4.18	2.85	1.00	18.00
Preceding Birth Interval				
< 18 months	0.43	0.50	0.00	1.00
18–23 months	0.21	0.40	0.00	1.00
24+ months	0.36	0.48	0.00	1.00
Neonatal Death of Prior Child (1 = yes)	0.02	0.15	0.00	1.00
Perinatal Death	0.02	0.14	0.00	1.00
Neonatal Death	0.00	0.03	0.00	1.00
Postneonatal Death	0.01	0.10	0.00	1.00
Died After First Year	0.97	0.17	0.00	1.00

*N = 1,777.

ence our estimates of the *total* effects of the variables we include.

FINDINGS

The weighted descriptive statistics for the variables in our analyses appear in Table 1. Our sample includes children of birth orders up to 18. Using the sample weights, we estimate that 2% of all infants died in the first week following birth, 0.10% died in the neonatal period, and 1% died in the post-neonatal period. The mean inbreeding coefficient of 0.0166 corresponds to inbreeding at the level of second cousins.

Table 2 presents distribution information for inbreeding by mortality component. Although the mean for the inbreeding coefficient is highest during the neonatal period (0.0224), there are only 16 neonatal deaths. Infants who died during the perinatal period have the second highest mean inbreeding coefficient (0.0174). However, no mortality group indicates an average inbreeding score below the level of second cousins.

Tables 3–5 present regression coefficients from our estimation models for the three mortality components. In

TABLE 2. DESCRIPTIVE STATISTICS FOR INBREEDING COEFFICIENT BY MORTALITY COMPONENT, EXCLUDING FIRSTBORN

Mortality Component	N	Mean	Standard Deviation	Minimum	Maximum
Perinatal	372	0.0174	0.0032	0.0000	0.0602
Neonatal	16	0.0224	0.0047	0.0051	0.0602
Postneonatal	150	0.0173	0.0035	0.0000	0.0602
Beyond First Year	1,239	0.0166	0.0114	0.0000	0.0592
Sibling Died (Neonatally)					
No	1,696	0.0166	0.0099	0.0000	0.0602
Yes	81	0.0165	0.0056	0.0000	0.0420

each of the three tables, Model 1 presents the coefficient of inbreeding without any controls. In Model 2, we exclude inbreeding to examine the total effects of the social and cultural variables and gender. Model 3 adds inbreeding to Model 2; we look for evidence of whether inbreeding changes the magnitude or direction of the coefficients. In Model 4, we include the proximate measures of birth order, birth interval, and maternal age; in Model 5, we introduce death of the preceding child. Model 6 shows whether coefficients for any variables change with the exclusion of inbreeding.

Table 3 presents the logistic regression estimates for perinatal mortality. None of the coefficients for inbreeding are statistically significant at the conventional 0.05 level.

Females have a lower perinatal mortality risk than do males (Models 5 and 6). Compared with childbirth for mothers over age 34, childbirth for mothers ages 21–34 significantly reduces the likelihood of perinatal death. A child whose sibling dies, however, has an increased chance of perinatal death compared with children whose sibling survives. Similarly, a birth interval of less than 18 months, compared with intervals of 24 months or longer, increases the likelihood of a perinatal death. The pattern of effects for these measures is similar across models. Excluding inbreeding in Model 6 produces no noteworthy changes in the direction or magnitude of coefficients for any covariates.

As shown in Table 4, inbreeding increases the chance of neonatal death, unlike the case for perinatal mortality. No other covariates emerge as significant predictors. There are a small number of neonatal deaths in our sample, however.

In the postneonatal models shown in Table 5a, the increased net risk from inbreeding is apparent; its absence suggests minimal influence on the effects of other measures in our models. Additional mortality risks include gender, maternal age, and birth order. Females face a greater likelihood of dying in the postneonatal period than do males, in contrast to the increased peril for males during the perinatal period. The influence of gender, however, is mediated by the proximate determinants in Model 4. Compared with those born to older mothers, postneonates whose mothers are under age 35 are at greater risk of death. Higher birth orders add another threat during this life stage.

Table 5a also reveals that children born more recently encounter a reduced chance of dying postneonatally relative to those born earlier. Living in a northern church district, which represents less traditional Amish culture, reduces the

TABLE 3. LOGISTIC REGRESSION COEFFICIENTS FOR PERINATAL MORTALITY

Independent Variable	Model 1	Model 2	Model 3	Model 4	Model 5	Model 6
Intercept	-1.43****	-1.05****	-1.24****	-1.36****	-1.35****	-1.19***
Inbreeding Coefficient*	0.06	—	0.11	0.11	0.10	—
Church District						
Northern	—	-0.15	-0.15	-0.12	-0.10	-0.10
Other	—	-0.18	-0.12	-0.12	-0.13	-0.19
Female (1 = yes)	—	-0.18	-0.19	-0.21	-0.26*	-0.25*
Date of Birth	—	-0.02****	-0.02****	-0.01****	-0.01****	-0.01***
Mother's Age						
< 21 years	—	—	—	-0.31	-0.38	-0.34
21–34 years	—	—	—	-0.41	-0.47*	-0.45*
First-Born Child	—	—	—	-0.11	0.04	0.03
Birth Order	—	—	—	0.02	0.01	0.01
Preceding Birth Interval						
< 18 months	—	—	—	0.62****	0.56****	0.56***
18–23 months	—	—	—	0.30	0.30	0.30
Preceding Sibling Died (1 = yes)	—	—	—	—	1.46****	1.47****

*Inbreeding coefficients are divided by 100.

* $p \leq .05$; *** $p \leq .001$; **** $p \leq .0001$

TABLE 4. LOGISTIC REGRESSION COEFFICIENTS FOR NEONATAL MORTALITY

Independent Variable	Model 1	Model 2	Model 3	Model 4	Model 5	Model 6
Intercept	-5.42****	-4.26****	-5.45****	-5.80**	-5.86**	-4.74**
Inbreeding Coefficient*	0.49**	—	0.60**	0.57**	0.58**	—
Church District						
Northern	—	-0.24	-0.21	-0.13	-0.14	-0.20
Other	—	0.33	0.66	0.71	0.70	0.32
Female (1 = yes)	—	-0.14	-0.18	-0.13	-0.15	-0.14
Date of Birth	—	-0.02	-0.03	-0.03	-0.02	-0.02
Mother's Age						
< 21 years	—	—	—	1.95	1.95	2.09
21-34 years	—	—	—	0.49	0.48	0.54
First-Born Child	—	—	—	-0.04	0.05	0.03
Birth Order	—	—	—	-0.03	-0.03	-0.04
Preceding Birth Interval						
< 18 months	—	—	—	-0.46	-0.50	-0.47
18-23 months	—	—	—	-0.08	-0.06	-0.06
Preceding Sibling Died (1 = yes)	—	—	—	—	1.08	1.01

*Inbreeding coefficients are divided by 100.

** $p \leq .01$; **** $p \leq .0001$

chances of death compared with living in the more traditional southern districts.

Table 5b presents regression coefficients for postneonatal mortality from our estimation models of Tables 3-5, but with the sample restrictions used by Curtis et al. (1993). Curtis et al. (1993) do not find statistically significant effects of birth order in their analysis. However, when we ex-

cluded birth order, a statistically significant coefficient in Table 5b, we obtained nonsignificant coefficients for maternal age 21-34 and reduced the level of significance for the inbreeding coefficient (data not shown). Therefore, we retained the birth order variable.

Unlike coefficients in Table 5a, our measures for date of birth and young maternal age (under 21) are not significant

TABLE 5A. LOGISTIC REGRESSION COEFFICIENTS FOR POSTNEONATAL MORTALITY

Independent Variable	Model 1	Model 2	Model 3	Model 4	Model 5	Model 6
Intercept	-2.42****	-1.86****	-2.33****	-4.00****	-3.97****	-3.47****
Inbreeding Coefficient*	0.18**	—	0.27**	0.29***	0.29***	—
Church District						
Northern	—	-0.54**	-0.54**	-0.50*	-0.50*	-0.50*
Other	—	-0.29	-0.16	-0.18	-0.18	-0.32
Female (1 = yes)	—	0.37*	0.35*	0.33	0.32	0.34
Date of Birth	—	-0.02****	-0.03****	-0.03****	-0.03****	-0.02****
Mother's Age						
< 21 years	—	—	—	1.29**	1.27*	1.35**
21-34 years	—	—	—	0.93**	0.91*	0.93**
First-Born Child	—	—	—	0.18	0.24	0.22
Birth Order	—	—	—	0.17****	0.17****	0.17****
Preceding Birth Interval						
< 18 months	—	—	—	-0.16	-0.18	-0.17
18-23 months	—	—	—	0.08	0.11	0.10
Preceding Sibling Died (1 = yes)	—	—	—	—	0.79	0.78

*Inbreeding coefficients are divided by 100.

* $p \leq .05$; ** $p \leq .01$; *** $p \leq .001$; **** $p \leq .0001$

TABLE 5b. LOGISTIC REGRESSION COEFFICIENTS FOR POSTNEONATAL MORTALITY, WITH SAMPLE RESTRICTIONS

Independent Variable	Model 1	Model 2	Model 3	Model 4	Model 5	Model 6
Intercept	-2.64****	-1.96****	-2.40****	-4.27****	-4.30****	-3.89****
Inbreeding Coefficient*	0.24*	—	0.26*	0.25*	0.25*	—
Church District						
Northern	—	-0.92**	-0.90**	-0.85**	-0.84**	-0.86**
Other	—	-0.29	-0.15	-0.17	-0.14	-0.28
Female (1 = yes)	—	0.49*	0.47	0.46	0.46	0.47
Date of Birth	—	0.00	-0.01	-0.01	-0.01	-0.01
Mother's Age						
< 21 years	—	—	—	1.52	1.56	1.69
21–34 years	—	—	—	1.02*	1.01*	1.04*
Birth Order	—	—	—	0.18**	0.18**	0.18**
Preceding Birth Interval						
< 18 months	—	—	—	0.14	0.08	0.09
18–23 months	—	—	—	0.16	0.18	0.17
Preceding Sibling Died (1 = yes)	—	—	—	—	0.84	0.81

*Inbreeding coefficients are divided by 100.

* $p \leq .05$; ** $p \leq .01$; **** $p \leq .0001$

in the models shown in Table 5b. Gender is significant only in Model 2 in Table 5b.

DISCUSSION

Using data from the Lancaster Old Order Amish settlement, we find that inbreeding, primarily a biological factor, increases the likelihood of death in the neonatal and postneonatal periods of early childhood, net of survival status of the immediately preceding sibling and other variables. The effect of inbreeding supports a differential familial mortality risk explanation: The more consanguineous the marriage, the greater the chances of dying during the first year for infants who survive the first week following birth.

In the perinatal models, the risk posed by inbreeding is statistically insignificant. Adding a product term (inbreeding by first-born child) in preliminary analysis did not produce a statistically significant effect for perinatal mortality. Perhaps the inbreeding effect is suppressed by other factors, such as prematurity and low birth weight (Khouri and Cohen 1987; Miller 1989).⁸ The deleterious effects from inbred families may not arise until the child has passed through the critical days after birthing. Our lack of perinatal effects could be due to past inbreeding among the Amish, eliminating deleterious recessive conditions and producing a genetic load that is milder and operates at slightly older ages.⁹ We cannot accept or refute these possibilities with the available data.

8. Khouri and Cohen (1987) report that the most common cause of infant death in the Lancaster settlement is prematurity, which increases the risk of prematurity in a subsequent pregnancy. They find that inbreeding does not directly affect prematurity, but increases the risks of congenital malformations, a lower intrauterine growth rate, and birth complications such as fetal distress (cf. McKusick et al. 1978).

9. We thank an anonymous reviewer for noting this possibility.

An alternative explanation for our perinatal results is a reporting bias of deaths, especially for deaths occurring immediately after a birth. Some underreporting of deaths is possible in the Amish directory, especially for earlier generations and for births occurring outside a hospital setting. Although we have no reliable information about possible omissions, an epidemiological study of mortality in the Lancaster settlement found that more than 90% of their recorded cases had death vital records (including fetal death) filed with the state health department (Khouri et al. 1987b). Although we cannot rule out underreporting effects, if severely affected infants who died immediately after birth were classified as stillborn (and not reported, or reported but excluded from analysis due to our sampling of live-born infants) and these infants were highly inbred, our results would underestimate the effect of inbreeding.

For neonatal mortality, our measure of inbreeding produced a statistically significant effect. Because there were few neonatal deaths in our sample, the results from these estimations are tentative.

Inbreeding clearly poses a significant threat of postneonatal death. Additional risks are incurred at higher birth orders and for infants of mothers under age 35. Residing in a northern church district reduces the chance of dying, perhaps because women living in northern districts are less reluctant than women in southern districts to seek prenatal and well-baby care. We do not find an increased likelihood of death from very short birth intervals, as Curtis et al. (1993) report, and we do not replicate their interactions between gender and maternal age. Our gender-mortality patterns are similar to those identified by Trussell and Hammerslough (1983), who find a lower mortality rate for females than for males that disappears around 6 months. Trussell and Hammerslough suggest that this pattern is due to more advantageous care

and feeding of males. Although the Amish is a traditional agricultural population, whether the Amish favor males in home and health practices is unclear. Preliminary analysis did not reveal statistical interaction effects between inbreeding and these variables, and we cannot search exhaustively here for specific explanations. Nonetheless, it is plausible that, in addition to an intrafamilial factor, inbreeding is linked to population-based social arrangements that influence postneonatal survival (e.g., access to and preference for medical care and the pace of fertility) and vary by district, over time, and perhaps by child's gender.¹⁰

In summary, we find that inbreeding increases the risk of neonatal and postneonatal mortality. Yet, the findings of the present study also reaffirm the relevance of incorporating social, demographic, and cultural measures in studies of child survival. We find clear evidence of the impact of these measures, even in a population in which biological influences are expected. It is well documented that the Lancaster Amish are likely to experience certain genetically transmitted defects that could increase the likelihood of mortality. Additional study is underway to clarify the specific pathways by which social, demographic, and cultural influences affect infant survival in this population, including the genetic risks associated with inbreeding across mortality components. We look forward to additional research examining familial and population-based mortality risks across a variety of consanguineous communities and under appropriate statistical controls.

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10. We are grateful to a reviewer of an earlier version of this paper suggesting this interpretation.

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